Case 16726

Eurorad

Acute hyperammonaemic encephalopathy after TIPS

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Section: Neuroradiology
Area of Interest: CNS
Procedure: Shunts
Imaging Technique: MR
Special Focus: Metabolic disorders Case Type: Clinical Cases
Authors: Arnaud Braeckman, Eva Genbrugge
Patient: 71 years, male

Clinical History:

A 71-year-old man with significant history of alcoholic liver cirrhosis with portal hypertension presenting at the emergency department with profuse haematemesis. Transjugular intrahepatic portosystemic shunt (TIPS) was performed. In the following 3 days, the patient deteriorated neurologically with reduced level of consciousness and seizures. Lab tests (hyperammonaemia 152µmol/L (5-55 µmol/L)) and MRI were performed to investigate the altered mental state. MRI was obtained 6 days after the first epileptic event.

Imaging Findings:

Figure 1: Axial T2-FLAIR image of the brain shows demarcated symmetrical swelling and oedema of the insular cortex, cingulate gyrus and medial frontal cortex, as well as signal abnormalities in both caudate nuclei and thalami. (Fig. 1)

Figure 2: Axial T2-FLAIR image of the brain on a lower level shows demarcated symmetrical swelling and oedema of the insular temporal cortex and hippocampi and sparing of the occipital cortex. (Fig. 2)

Figure 3: Diffusion-weighted image (a) and ADC map (b) shows clear signal hyperintensity in the swollen cortical zones. The ADC map shows concordant hypointensities, confirming the restricted diffusion. (Fig. 3a, b)

Figure 4: Follow-up T2-FLAIR image of the brain after 15 days at the same level as Fig. 1, comparison shows decreased oedematous aspect of the affected cortex.

Figure 5: Follow-up DWI (a) and ADC (b) of the brain after 15 days at the same level as Fig. 3, comparison shows decreased oedematous aspect of the affected cortex.

Discussion:

1. Background

Acute hyperammonaemic encephalopathy (AHAE) or portal-systemic encephalopathy represents a potentially reversible clinical syndrome during acute liver disease with impairment of neuropsychiatric function. Due to portosystemic shunting, there is no metabolisation of ammonia in the liver. Although the exact mechanism of neurotoxicity is not known, the uptake of ammonia by the brain has been shown to be toxic to astrocytes. It is not known why the insular and cingulate cortices seem particularly susceptible to the toxic effects of hyperammonaemia. [1] This case shows a possible side effect of TIPS placement. Once the TIPS is placed, toxin-rich blood bypasses the liver via the portosystemic shunt. This will cause an acute rise of many waste product blood levels like ammonia which will no longer be broken down into urea. Together with the excessive nitrogen-containing intestinal load following the recent GI bleeding, it results in an even higher ammonia blood level.

2. Clinical Perspective

Symptoms of AHAE range from changes in behaviour, mild confusion, slurred speech and disordered sleep to coma and unresponsiveness to pain. Because of the non-specific nature of these symptoms, additional lab tests and imaging are required to narrow down the differential diagnosis. High blood ammonia levels (above 55 µmol/L) and specific patterns of imaging findings should alert the clinician. [1] Subsequently, it is of great importance that radiologists recognise this pattern and report the likely association with hyperammonaemia.

3. Imaging Perspective

Key findings in AHAE are bilateral symmetrical T2/FLAIR hyperintense signal in the cortex of the insula and cingulate gyrus, and sparing of the perirolandic and occipital cortex. There can be a varying extension and degree of symmetry in the affected frontal, temporal and parietal cortices. Signal abnormalities in the thalami and globi pallidi are also a frequent finding, indicative of a pre-existing chronic disease. [2-4]
Proton-MR-spectroscopy can be performed when available and will show an increase in the glutamate/glutamine resonance and a decrease in the myo-inositol and choline resonances. [5]

4. Outcome

Findings show that the cortical changes appear to be early and reversible findings, if aggressive treatment is instituted. Probably the plasma ammonium levels may reflect the extent of the injury. Commonly used treatments for hepatic encephalopathy aim to reduce ammonia production and absorption. This is accomplished by correcting hypokalaemia, giving synthetic disaccharides (such as lactulose) and/or antibiotics, and favouring colonisation with non-urease-producing bacteria. Or in this case, if medication doesn't work, reducing the TIPS. Patients with hepatic encephalopathy may have persistent and cumulative neurologic deficits despite an apparent normalisation of mental status after treatment (deficits in working memory, response inhibition, and learning when assessed by psychometric testing). [6]
Follow-up MRI was performed 15 days after the first MRI-scan and showed marked decrease of cerebral oedema. In an effort to treat the encephalopathy even more, the TIPS was occluded endovascularly. Unfortunately the patient in this case died due to recurrent GI bleeding.

5. Take Home Message / Teaching Points

Symmetrical cortical signal abnormalities in the insular cortex and cingulate gyrus with sparing of the perirolandic and occipital cortices are key findings in MRI imaging of AHAE. The diagnosis should be suggested particularly if there is a recent history of TIPS placement.

Written informed patient consent for publication has been obtained.

Differential Diagnosis List: Acute hyperammonaemic encephalopathy after TIPS, Hypoxic-ischaemic brain injury, Acute hyperammonaemic encephalopathy, Uraemic encephalopathy, Valproate-induced hyperammonaemia, Adult-onset citrullinaemia, Reye syndrome, Creutzfeld-Jakob disease

Final Diagnosis: Acute hyperammonaemic encephalopathy after TIPS

References:

Vilstrup H, Amodio P, Bajaj J, Cordoba J, Ferenci P, Mullen KD, Weissenborn K and Wong P (2014) Hepatic encephalopathy in chronic liver disease: 2014 Practice Guideline by the American Association for the Study of Liver Diseases and the European Association for the Study of the Liver. 60: 715-735 (PMID: 25042402)
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McKinney AM, Kieffer SA, Paylor RT, SantaCruz KS, Kendi A, Lucato L (2009) Acute Toxic Leukoencepha¬lopathy: Potential for Reversibility Clinically and on MRI With Diffusion-Weighted and FLAIR Imag¬ing. Am J Roentgenol 193(1): 192–206 (PMID: 19542414)
U-King-Im JM, Yu E, Bartlett E, Soobrah R, Kucharczyk W (2011) Acute hyperammonemic encephalopathy in adults: imaging findings. American journal of neuroradiology 32 (2): 413-8 (PMID: 21087942)
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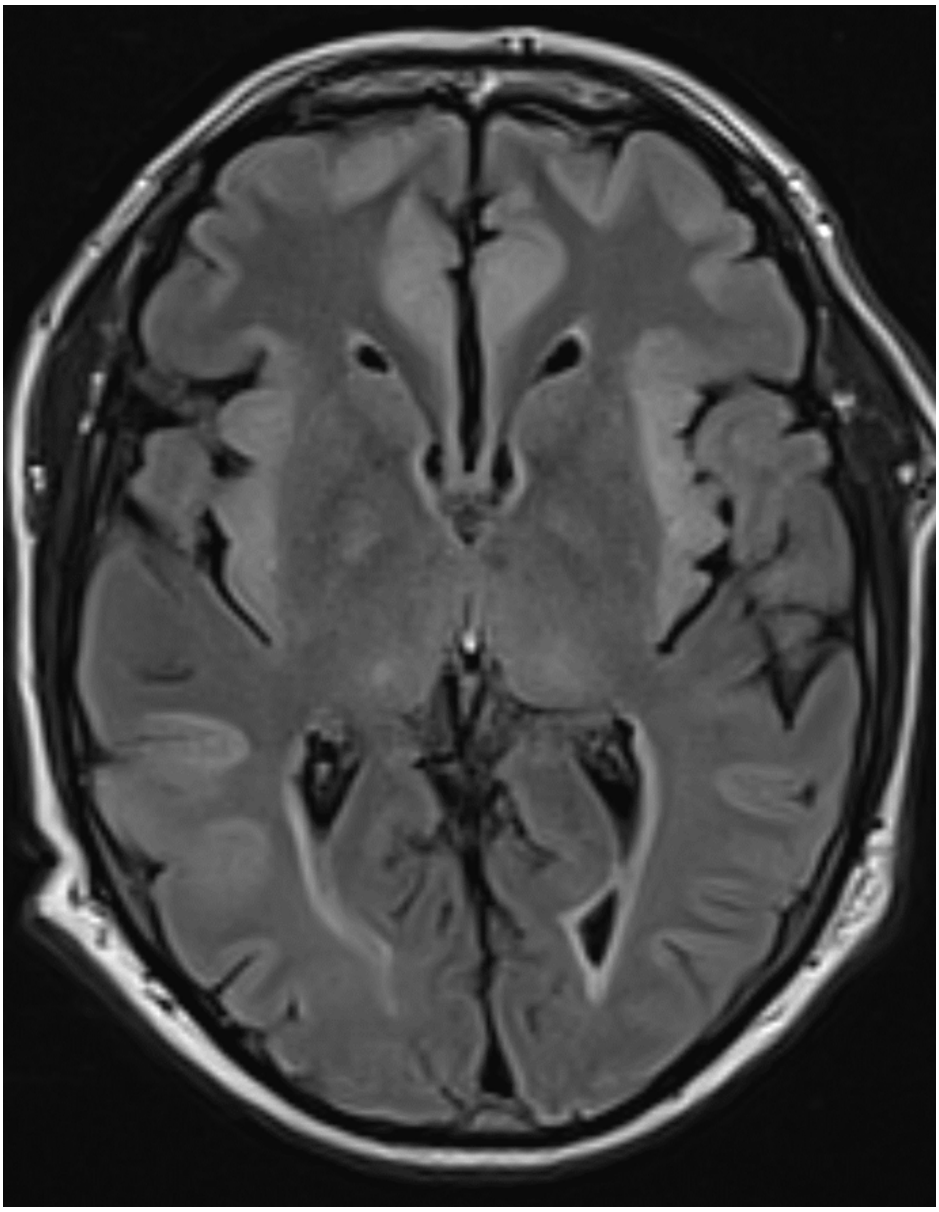
Bajaj JS, Schubert CM, Heuman DM, et al (2010) Persistence of cognitive impairment after resolution of overt hepatic encephalopathy. Gastroenterology 138(7): 2332-2340 (PMID: 20178797)

Figure 1

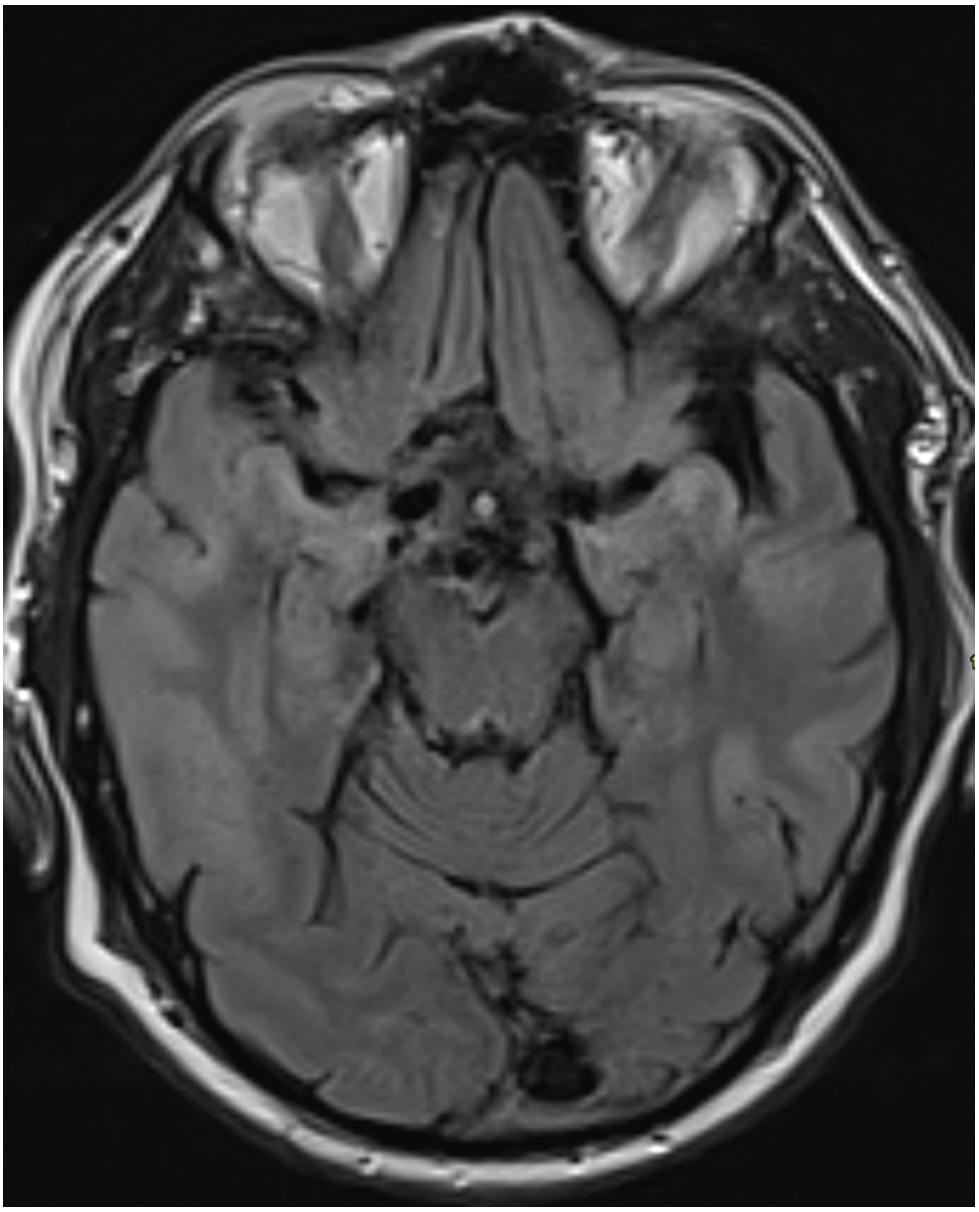
a



Description: Axial T2-FLAIR image of the brain shows demarcated symmetrical swelling and oedema of the insular cortex, cingulate gyrus and medial frontal cortex, as well as signal abnormalities in both caudate nuclei and thalami. (Fig. 1) **Origin:** Radiology Department, University Hospital of Ghent (UZGent)

Figure 2

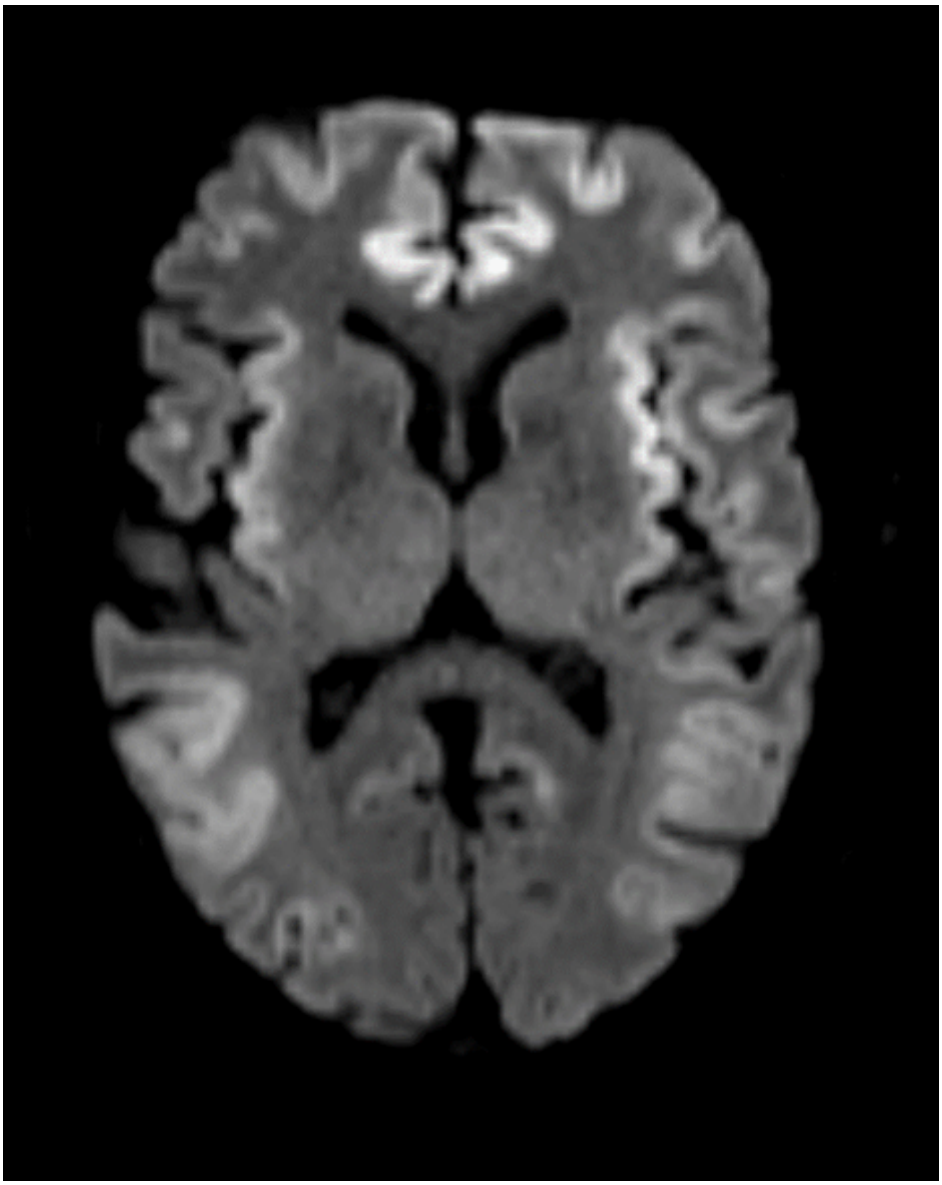
a



Description: Axial T2-FLAIR image of the brain on a lower level shows demarcated symmetrical swelling and oedema of the insular temporal cortex and hippocampi and sparing of the occipital cortex. (Fig. 2) **Origin:** Radiology Department, University Hospital of Ghent (UZGent)

Figure 3

a



Description: Diffusion-weighted image (a) and ADC map (b) shows clear signal hyperintensity in the swollen cortical zones. The ADC map shows concordant hypointensities, confirming the restricted diffusion. (Fig. 3a, b) **Origin:** Radiology Department, University Hospital of Ghent (UZGent)

b

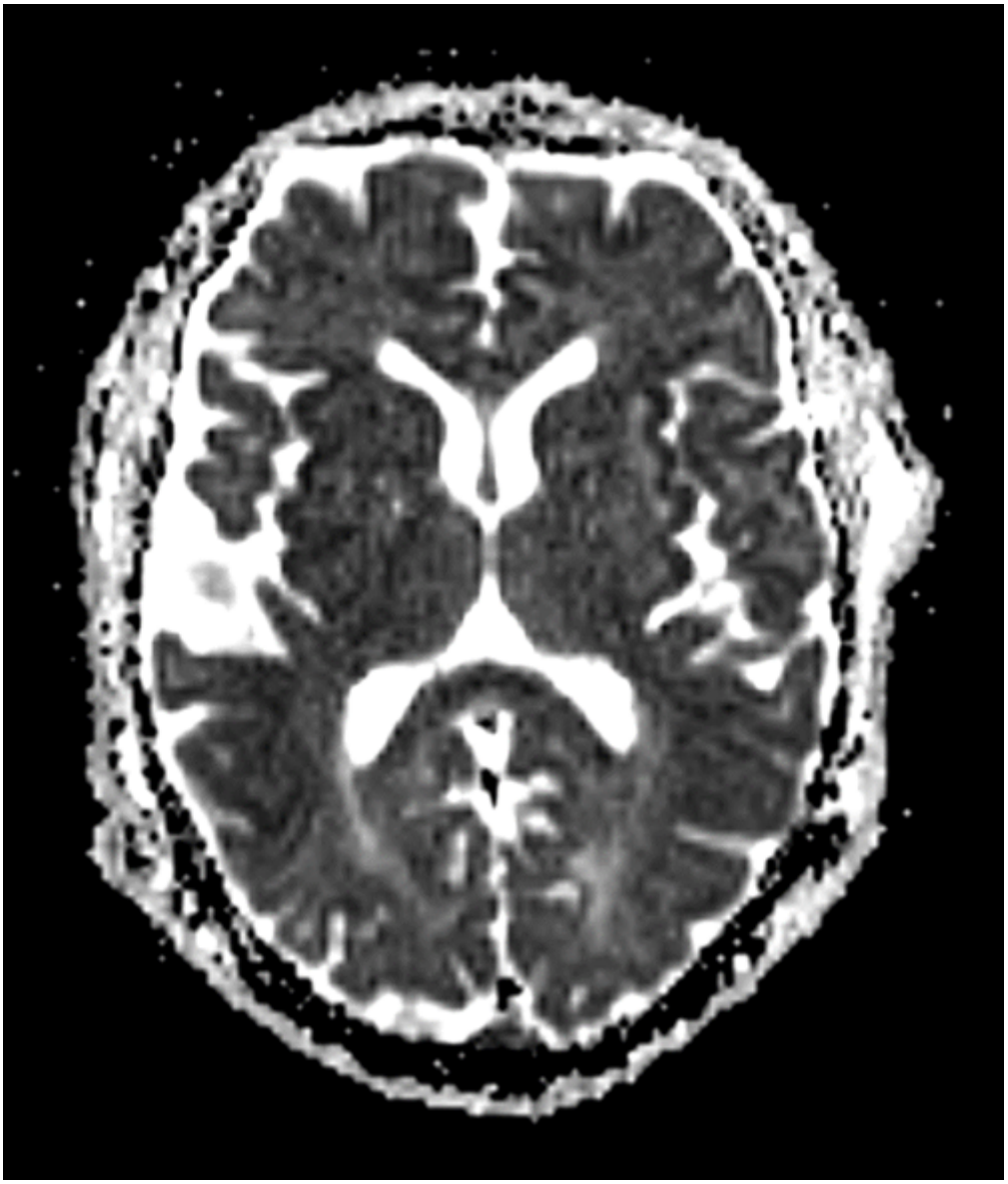


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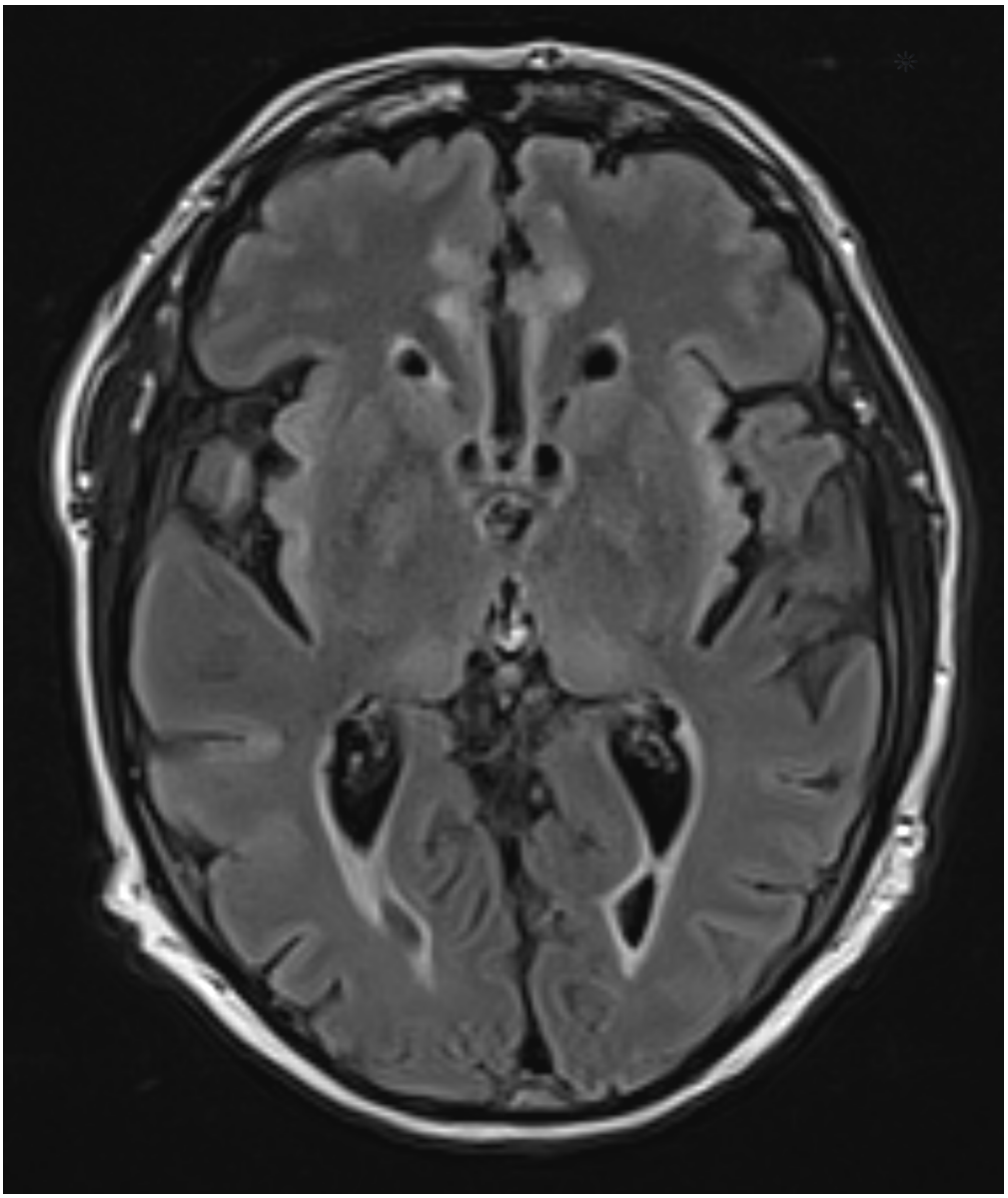
Figure 4

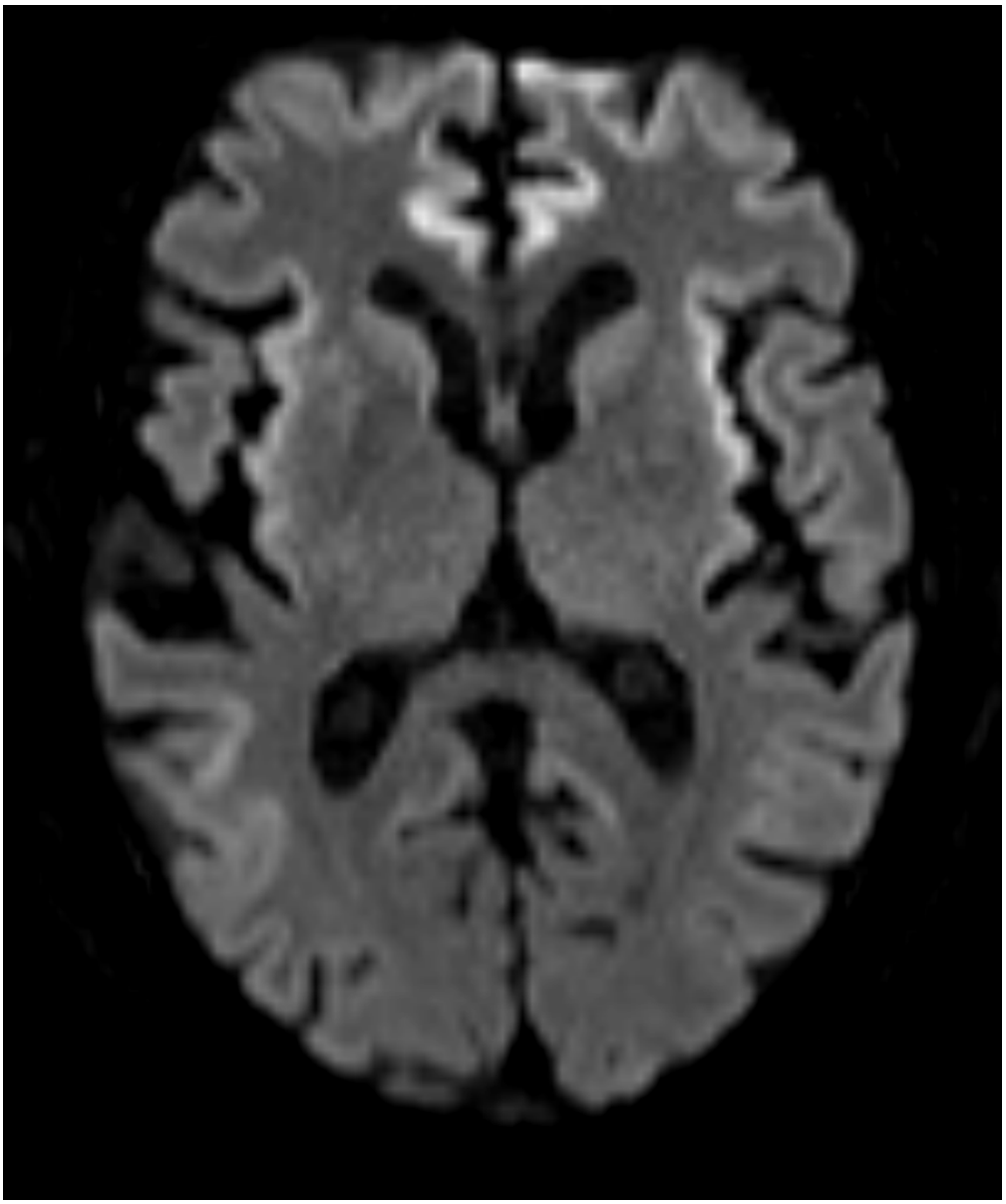
a



Description: Follow-up T2-FLAIR image of the brain after 15 days at the same level as Fig. 1, comparison shows decreased oedematous aspect of the affected cortex. **Origin:** Radiology Department, University Hospital of Ghent (UZGent)

Figure 5

a



Description: Follow-up DWI (a) and ADC (b) of the brain after 15 days at the same level as Fig. 3, comparison shows decreased oedematous aspect of the affected cortex. **Origin:** Radiology Department, University Hospital of Ghent (UZGent)

b

Description: Follow-up DWI (a) and ADC (b) of the brain after 15 days at the same level as Fig. 3, comparison shows decreased oedematous aspect of the affected cortex. **Origin:** Radiology Department, University Hospital of Ghent (UZGent)